ANGIOEDEMA AND HEREDITARY ANGIOEDEMA

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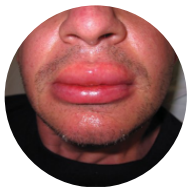
ANGIOEDEMA

- Angioedema is a rare problem with episodes of swelling that occurs mostly in the hands and feet, abdomen, face, genitals and throat.
- The swelling is more raised than hives, less red, and firm.
- Angioedema may come and go over a longer period than hives and last for up to 3 days.
- Angioedema is not itchy but may be painful.
- If angioedema occurs along with an itchy, red or pink rash ("urticaria," hives, "bommels" or wheals) that has a round or wavy border and comes and goes over minutes to hours leaving no trace on the skin behind, it is often caused by an allergy (see urticaria).
- However if angioedema occurs on its own, without urticaria, it is either caused by specific medication (ACE inhibitors) or because of a genetic problem called hereditary angioedema.

DRUG (ACE-INHIBITOR) RELATED ANGIOEDEMA



- Angioedema affects only very few patients taking ACE inhibitors.
- It can occur with the first time taking the drug or only after many years.
- Swelling can be very severe and may be life-threatening.
- It often involves the face, lips and eyes. Tongue and throat swelling can cause severe breathing difficulty.
- If this reaction occurs, the ACE inhibitor must be stopped immediately. Even if the angioedema goes away quickly, it may recur, so all patients should be admitted overnight.
- All types of ACE inhibitors must be avoided, as the reaction is caused by all of the related drugs.
- A "cousin" of ACE inhibitors, the "angiotensin II receptor antagonists" are almost always safe to use.
- Patients should wear a medic alert or similar bracelet.
- Patients should be screened for underlying hereditary angioedema as ACE reactions are more common in this group.

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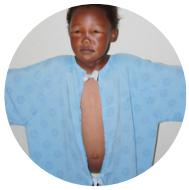
HEREDITORY ANGIOEDEMA (HAE)



- HAE is a genetic problem that runs in families.
- Patients have attacks of swelling that may affect any part of the body.
- It is not associated with hives but some patients may have a soft pink non-itchy rash before the angioedema starts.
- Swelling may occur in the lining of the gut, causing severe stomach pain and vomiting that is often mistaken for appendicitis or other problems.
- Swelling of the throat can cause difficult breathing and death.
- Within a single family some people can have mild attacks and others very severe attacks.

WHAT CAUSES HAE?

- HAE is called “hereditary” because the genetic problem is passed on in families. A child has a 50% chance of inheriting this disease if one of his or her parents has it.
- HAE can also occur for the first time in a person. Up to 25% of HAE cases are spontaneous genetic mutations and there is no family history. These patients can still pass the abnormal gene on to their children.
- HAE patients have a problem in the gene that controls a blood protein called C1 Inhibitor. The genetic problem causes low levels of C1-Inhibitor protein or a C1-Inhibitor protein that doesn’t work properly.
- Normal C1-Inhibitor helps to keep fluid within our blood vessels. If the inhibitor is missing, fluid leaks into the surrounding tissue causing angioedema.

TESTING FOR HAE

- Because the disease is very rare, patients often remain undiagnosed for many years.
- There are blood tests for HAE. If you have HAE, all members in the family should have the blood test to see whether they are at risk, even if they have never had any attacks.

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TREATMENT

Because HAE can be fatal, people with HAE must immediately recognise when they are having an attack and get medical help urgently.
Abdominal attacks and throat attacks are particularly dangerous.
Anyone with the slightest hint of throat involvement must urgently get extra treatment for an "acute attack".
Because it is a rare condition, doctors and nurses may not be familiar with HAE.
Patients must get an emergency plan (see pamphlet: HAE emergency plan) from their doctor and must wear a Medic Alert disc.
The emergency plan must be carried with them at all times to tell other doctors about their condition and the correct treatment.
Patients with HAE must be under the regular care of a doctor who has experience with the condition.

TREATMENT OF ACUTE ATTACKS

- The swelling of HAE can occur quickly and cause death. At the first sign of an attack, the progression of the swelling must be stopped, particularly if it affects the airway.
- Acute therapies currently available in South Africa include:
  - **Fresh-frozen plasma (FFP)** -given as an intravenous infusion in hospital.
  - **Firazyr (Icatibant)** – Indicated for treatment of acute attacks of HAE in patients from two years and older. Firazyr is delivered by subcutaneous injection and may be self-administered.
  - **Berinert** – indicated for the treatment of HAE attacks in adult and paediatric patients, delivered intravenously.
  - **Ruconest** – indicated for use in for treating HAE attacks in adults and adolescents, delivered intravenously.
  - **Berinert and Ruconest** are not currently registered for use in S.A. and can only be accessed via Section 21 application)
- The treatment usually given for hives is NOT effective.
- Other treatments include pain relief for abdominal attacks, medication to prevent vomiting and extra fluids.

SHORT-TERM PREVENTION

- Before surgery or dental work, short term treatment is needed to avoid the risk of any attacks.
- Prophylactic treatment may be used 1-12 hours before the surgery.
- In adults an oral medication (of "high-dose androgens") may be given for the previous 5–10 days.

LONG-TERM PREVENTION

- Patients in whom episodes occur at least once a month or who are at high risk of developing throat attacks need long-term prevention.
- Oral androgen hormone medications increase production of C1-INH in the liver but have some side effects particularly in women and children and they must not be taken during pregnancy.
- As an alternative, large doses tranexamic acid or aminocaproic acid can be used. The effect is weaker than androgens but side effects are less severe.

KEY POINTS

- If angioedema occurs on its own, without urticaria, it is usually caused by ACE inhibitors or hereditary angioedema.
- The correct diagnosis must be established.
- Your doctor will complete an application form for a Medic-Alert bracelet and an action plan detailing emergency treatment.
- Patients suffering from acute attacks must seek emergency help immediately.

A medical specialist with a special interest and skill in allergy might be able to help. See the list of health professionals with skills in allergy on the AFSA website: www.allergyfoundation.co.za

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HEREDITARY ANGIOEDEMA SUPPORT

Hereditary Angioedema South Africa Support:

Please contact Janice Strydom at haeadvocacysa@gmail.com or Tamsin Van Vlaanderen at haesouthafrica@gmail.com or visit their website https://southafrica.haei.org/

The "Sinovuyo" – South African Virtual Angioedema Centre offers person to person virtual consultations with angioedema specialists: https://aiu.haei.org/

Hereditary Angioedema International Support (HAEi)

For Hereditary Angioedema International Support kindly visit their website https://haei.org/